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CASE REPORT

Hirayama Disease with Asymmetric Bilateral Electrophysiological Involvement in a 14-Year-Old Male: A Case Report

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ABSTRACT

Hirayama disease is an uncommon cervical myelopathy, progressive at first and then self-limiting, caused by repetitive neck flexion, typically presenting in adolescent males as insidious wasting and weakness of the distal upper limb. We describe a 14-year-old boy with a six-month history of right-hand weakness, with progressive hand and forearm atrophy for four months (stable for the last month), accompanied by diminished grip strength but intact sensation. Electrophysiological testing revealed a reduced compound muscle action potential (CMAP) in the right ulnar nerve, and needle EMG identified a neurogenic pattern in the C6–T1 distribution on the right and, on the left, the abductor digiti minimi (C8–T1), indicating bilateral, asymmetric involvement. Flexion-triggered dynamic MRI of the cervical spine showed forward shifting of the posterior dura, engorgement of the epidural venous plexus, and cord compression — findings that together supported the diagnosis. Management consisted of a rigid cervical collar and physiotherapy (with B-complex vitamins). This case illustrates how combining EMG with dynamic MRI can be decisive for catching Hirayama disease early, even when a predominantly right-sided clinical picture is accompanied by bilateral electrophysiological involvement.

KEYWORDS

• Hirayama Disease • Cervical Flexion Myelopathy • Electromyography • Dynamic MRI • Anterior Horn Cell Disease

INTRODUCTION

First described by Hirayama and colleagues in 1959, Hirayama disease is understood today as a self-limiting form of cervical myelopathy, sometimes called juvenile muscular atrophy of the distal upper extremity.[1] It selectively damages the anterior horn cells spanning the C7–T1 cord segments and is seen predominantly in males during their teens and twenties.[2] The leading explanation for

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its mechanism centers on forward movement of the posterior dural sac each time the neck is flexed, which over time produces low-grade ischemic damage to the anterior horn.[3] Because the disease tends to plateau once neck movement is restricted, prompt diagnosis and cervical immobilization are critical to halting further deterioration. What makes the present case worth reporting is the mismatch between a clinical picture that looked predominantly right-sided and electrophysiological testing that told a more bilateral story.

CASE HISTORY

A 14-year-old boy was first seen at Jawaharlal Nehru Medical College (JNMC), Aligarh Muslim University, and later referred for further work-up to the Department of Neurology at Safdarjung Hospital, New Delhi. On admission (24 December 2025) he described a six-month history of trouble gripping objects with his right hand, along with visible thinning of the right hand and forearm over the preceding four months. The symptoms had crept in gradually rather than appearing suddenly. Everyday tasks had become difficult — buttoning his shirt, tearing roti, lifting a bucket of water, and opening bottle caps. In the month before he was admitted, the weakness had stopped worsening and stayed steady.

He denied any neck pain or trauma, sensory loss or tingling, twitching, bowel or bladder issues, cranial nerve symptoms, or preceding fever or viral infection. His personal history was unremarkable and no family member had a similar illness.

CLINICAL EXAMINATION

General Examination

He was alert and fully oriented, afebrile, with vitals within normal range (BP 113/84 mmHg, pulse 78/min, respiratory rate 16/min, SpO_2 98% on room air). There was no pallor, jaundice, cyanosis, clubbing, lymph node enlargement, or peripheral edema, and cardiac, respiratory, and abdominal exams were all unremarkable.

Neurological Examination

He was conscious and oriented. Cranial nerve examination was normal on both sides, with no cranial nerve involvement.

Motor Examination

Muscle wasting was visibly evident in the right hand and forearm (Figures 1 and 2), most notably in the intrinsic hand muscles, while tone remained normal across all limbs. Weakness clustered in the distal right upper limb — affecting the abductor pollicis brevis, flexor pollicis longus, flexor pollicis brevis, opponens pollicis, adductor pollicis, opponens and abductor digiti minimi, lumbricals, and the interossei — with subtler weakness of the opponens digiti minimi and abductor digiti minimi noted on the left as well. Grip strength was reduced on the right, with mild weakness of right elbow extension and wrist flexion/extension. Reflexes were symmetric and normal throughout, plantar responses were flexor bilaterally, and there were no upper motor neuron signs. Power in the right upper limb was 4/5 and in the left upper limb was 5/5 according to the MRC grading system. The brachioradialis muscle was spared.



Figure 1: Clinical photograph showing asymmetric wasting of the distal upper limb muscles, more pronounced on the right side, involving the hand and forearm.



Figure 2: Palmar view demonstrating asymmetrical wasting of the intrinsic muscles of the right hand with relative preservation of the left hand.

Sensory Examination

All sensory modalities — light touch, pain, temperature, vibration, and proprioception — were preserved on both sides. Romberg's test was negative and his gait was unremarkable. Taken together, the clinical picture pointed to an evolving lower motor neuron process affecting the distal upper limbs, right greater than left, with sensory pathways and corticospinal tract function unaffected.

Investigations

Routine bloodwork (14 July 2025) — complete blood count, liver and kidney function tests, electrolytes, and urinalysis — returned entirely normal.

Nerve Conduction Studies (NCS)

NCS were carried out on 26 December 2025 at the Clinical Neurophysiology Laboratory, VMMC & Safdarjung Hospital, New Delhi, testing the median, ulnar, common peroneal, tibial, and sural nerves bilaterally. The right ulnar nerve showed a reduced CMAP amplitude; sensory conduction was preserved across every nerve tested, and lower-limb studies were normal throughout — a pattern pointing toward isolated motor pathway involvement and anterior horn cell degeneration. An earlier study at JNMC, Aligarh (23 October 2025, right side only; Figure 3) had shown an increased right median distal latency, markedly reduced ulnar and radial CMAP amplitudes and mildly reduced sensory velocities; the sensory findings were normal on repeat testing at Safdarjung, the ulnar CMAP reduction persisted, and the radial nerve was not retested.

Electromyography (EMG)

Needle EMG (30 December 2025, Figure 4) sampled the right deltoid, triceps, first dorsal interosseous, abductor pollicis brevis, and extensor indicis proprius, plus the right vastus lateralis, tibialis anterior, and thoracic paraspinals, along with the left triceps, abductor digiti minimi and extensor indicis proprius; the left triceps and the sampled lower-limb and paraspinal muscles were normal. Chronic neurogenic changes were found in the C6–T1 distribution on the right and in the left abductor digiti minimi (C8–T1) — consistent with bilateral anterior horn cell involvement that was more pronounced on the right. The abnormal muscles were, on the right, the triceps, extensor indicis proprius, first dorsal interosseous and abductor pollicis brevis (polyphasic units with incomplete recruitment) and, on the left, the abductor digiti minimi (giant polyphasic units with incomplete recruitment); the left extensor indicis proprius and right deltoid were normal. The right deltoid (C5–C6) was spared, so the involvement was confined to the lower cervical myotomes, overlapping the C7–T1 myotomes classically affected in Hirayama disease.[3]

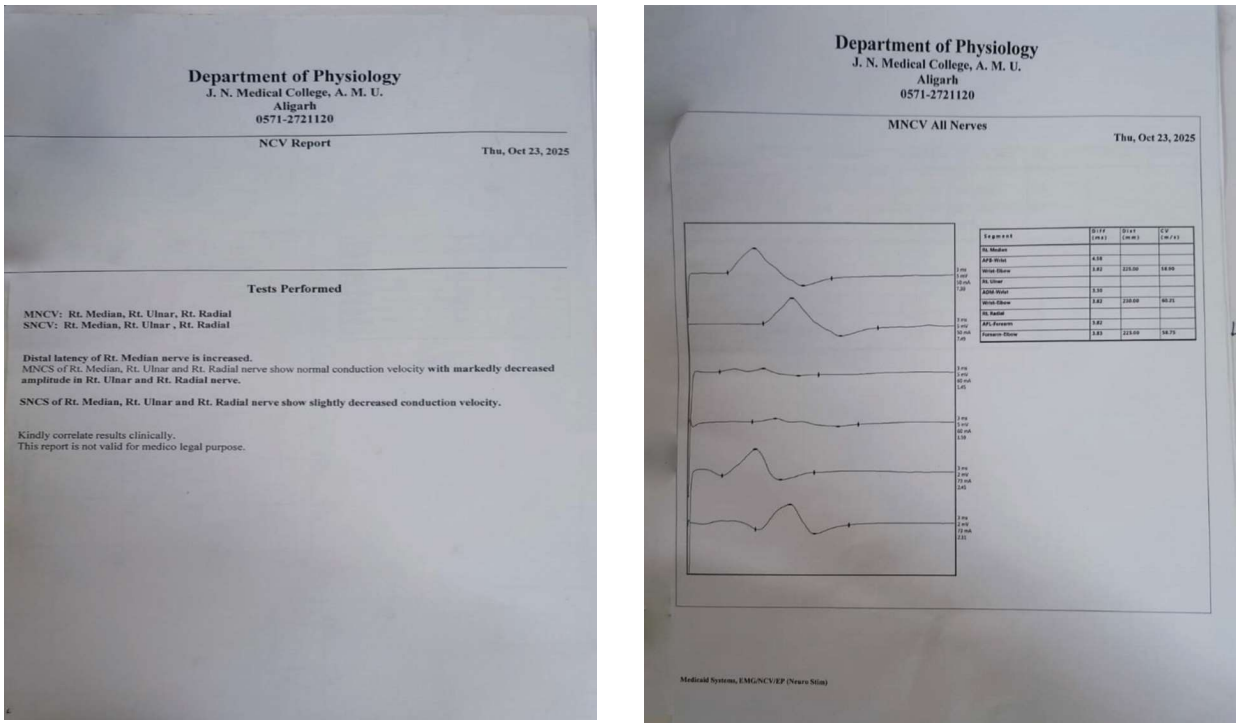


Figure 3: Nerve conduction study and electromyography demonstrating reduced CMAP amplitude in the right ulnar nerve and neurogenic changes involving the C6–T1 myotomes.

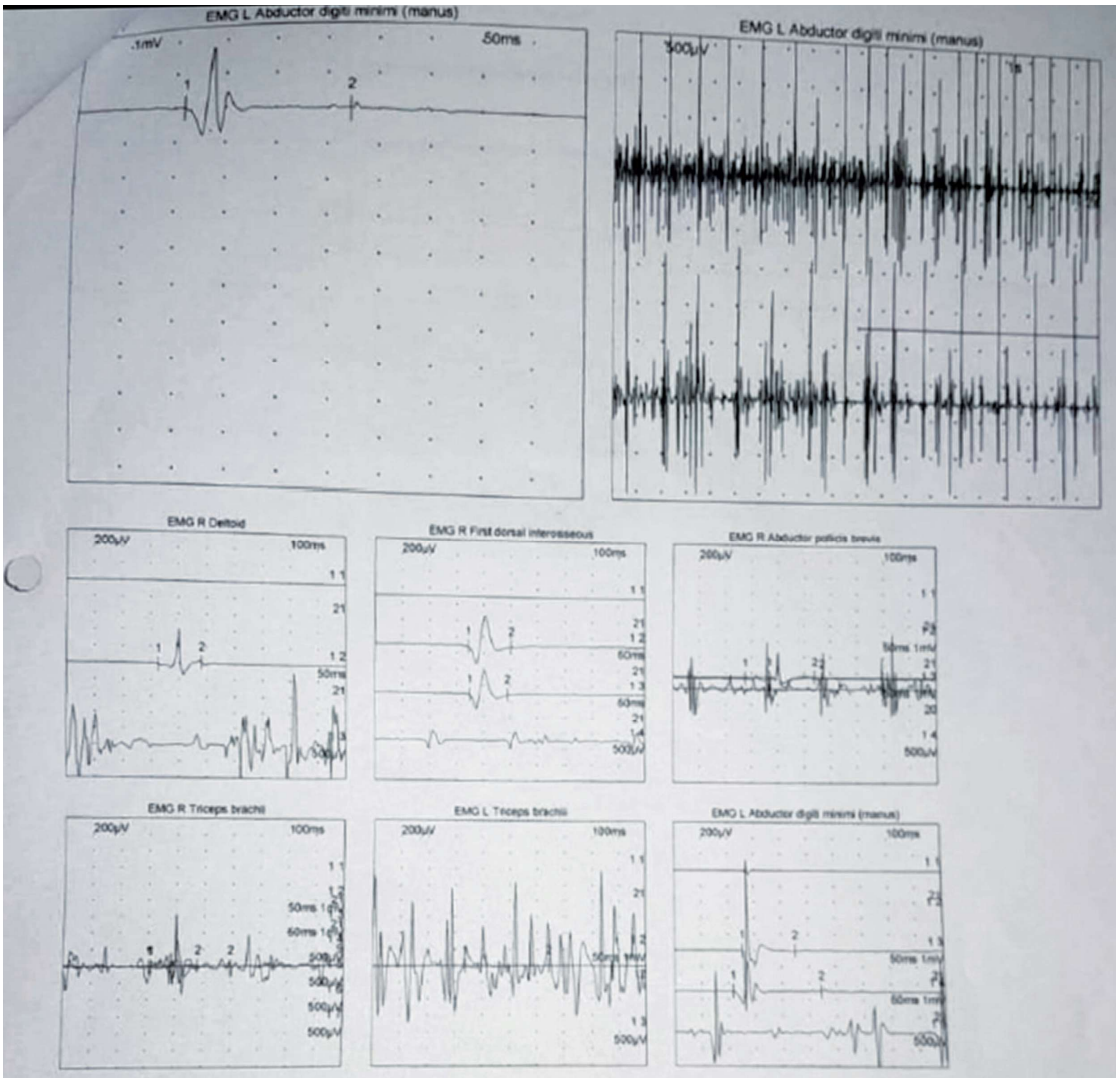


Figure 4: (Safdarjung Hospital): Needle EMG (30 Dec 2025) showing chronic neurogenic MUAP changes, including a large polyphasic unit in the left abductor digiti minimi and reduced recruitment in right hand muscles.

Neuroimaging

Brain MRI was unremarkable (outside centre, 27 October 2025). Dynamic contrast-enhanced cervical spine MRI performed in neutral and flexion positions (17 January 2026, performed as an outpatient after discharge; an earlier outside-centre flexion MRI on 31 October 2025 was reported as suggestive of Hirayama disease) showed straightening of the cervical spine. On flexion, the dorsal dura was detached and displaced anteriorly by a crescent-shaped epidural mass in the posterior epidural space from C4–C7, which was hyperintense on T2-weighted images, contained flow voids, and showed significant post-contrast enhancement, consistent with distension of the posterior internal vertebral venous plexus.[4] This caused severe flexion-related narrowing of the spinal canal (anteroposterior diameter approximately 5.2 mm, as stated in the radiology report) with compression and flattening of the cervical cord and intrinsic T2 hyperintensity within the cord — a combination of findings characteristic enough to support Hirayama disease (Figure 5).

Differential Diagnosis

Considerations included spinal muscular atrophy,[5] syringomyelia,[5] lower trunk brachial plexopathy,[5] cervical spondylotic amyotrophy,[6] and amyotrophic lateral sclerosis[6] — the last ruled out given the patient's age, lack of upper motor neuron signs, and the MRI findings.

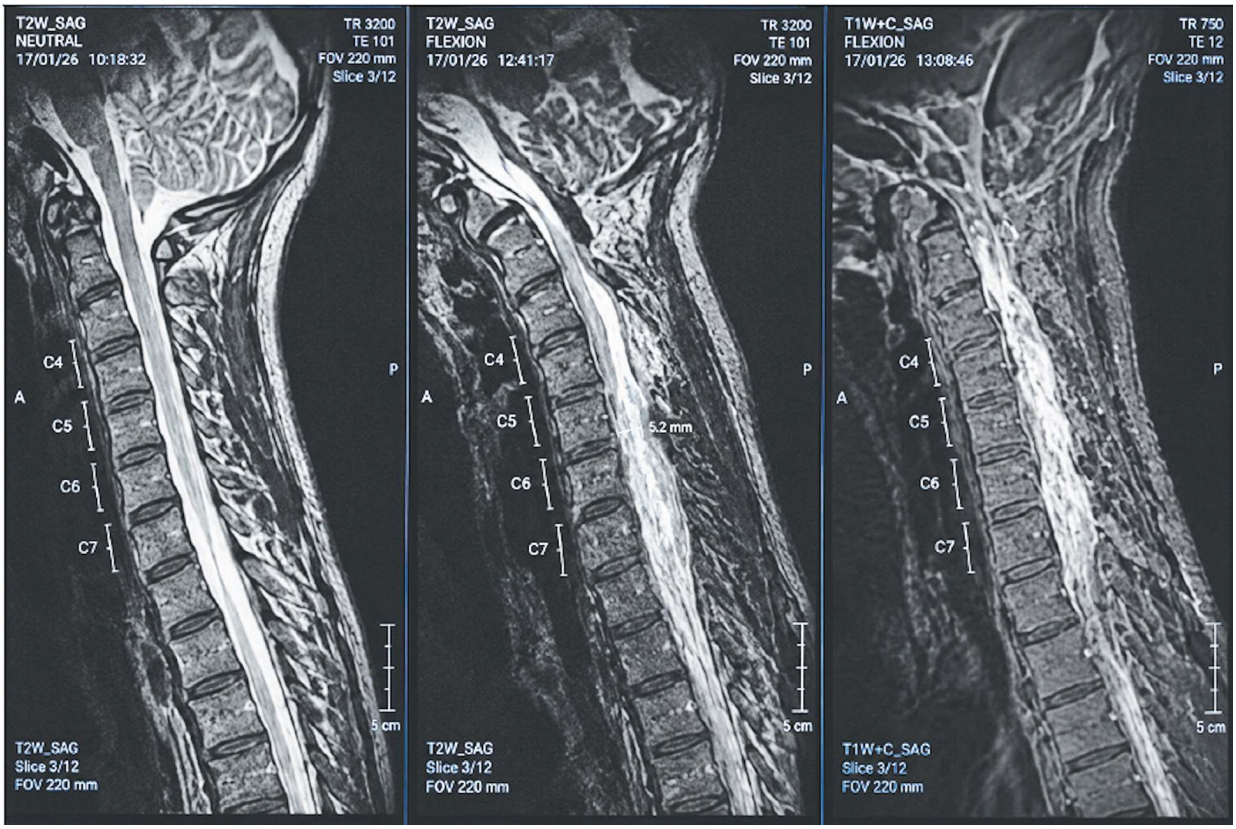


Figure 5: Cervical spine MRI, 17 January 2026. Left, T2-weighted sagittal image in neutral position. Middle, T2-weighted sagittal image in flexion. Right, T1-weighted post-contrast sagittal image in flexion. In flexion, there is a crescent-shaped posterior epidural collection from C4–C7 with anterior displacement of the dorsal dura, severe spinal canal narrowing, and cord compression with intrinsic T2 hyperintensity, in contrast to the neutral position. Vertebral levels C4–C7 are marked.

Management

Having first presented to JNMC, Aligarh Muslim University, he was subsequently admitted (24 December 2025) to the Neurology ward at Safdarjung Hospital, New Delhi. He was managed conservatively — a rigid cervical collar to limit flexion and offload the cord[7], a physiotherapy regimen to preserve muscle bulk and prevent contractures, and oral B-complex vitamins for 7 days to support nerve health. He was discharged on 1 January 2026 with instructions to keep wearing the collar, continue his physiotherapy routine, and return after 14 days to the Neurology outpatient clinic with the dynamic cervical MRI report for reassessment; the dynamic MRI was arranged after discharge and performed on 17 January 2026.

DISCUSSION

Hirayama disease is an uncommon, flexion-related cervical myelopathy that mainly affects adolescent and young adult men. It typically presents with a slow, painless wasting and weakening of the hand and forearm, sparing of sensation, and no upper motor neuron findings. The prevailing theory attributes it to repeated cord compression during neck flexion, which produces chronic ischemic injury to the anterior horn cells over time.

This patient's presentation — painless, progressive right-sided hand and forearm wasting with intact sensation and normal reflexes — fit the textbook picture closely, and the mild contralateral involvement placed him within the asymmetric bilateral subtype described in prior literature[8].

Electrophysiology was central to reaching the diagnosis here. Reduced CMAP amplitude in the right ulnar nerve alongside preserved sensory responses on repeat testing pointed to a pure motor process, while EMG uncovered chronic neurogenic changes bilaterally (C6–T1 on the right; left abductor digiti minimi, C8–T1) — a bilateral electrophysiological signature that has been documented before even in patients whose symptoms appear one-sided clinically.[8]

Dynamic contrast-enhanced MRI strongly supported the diagnosis, capturing the anterior shift of the posterior dura during flexion, an enhancing, congested epidural venous space from C4–C7, cord flattening, and T2 signal change within the cord — a constellation that reflects the flexion-driven compression and venous congestion underlying this disease.

Because restricting neck flexion can arrest further decline, early recognition matters. This patient was managed conservatively with a cervical collar and physiotherapy, and follow-up is planned to assess his response. The case reinforces two points worth emphasizing: first, that Hirayama disease can meaningfully disrupt an adolescent's daily function during a formative period of life, and second, that dynamic MRI paired with bilateral electrophysiological testing is especially valuable when the clinical picture looks asymmetric.

CONCLUSION

This report describes a 14-year-old boy whose progressive distal upper limb wasting, preserved sensation, and absent upper motor neuron signs were characteristic of Hirayama disease, even though electrophysiology was consistent with anterior horn cell involvement on both sides, in an outwardly predominantly right-sided presentation. Dynamic contrast-enhanced MRI strongly supported the diagnosis by demonstrating flexion-induced anterior dural displacement, posterior epidural venous engorgement, cord compression, and intramedullary signal changes. As this case shows, catching Hirayama disease early and starting cervical immobilization promptly are believed to protect neurological function over the long run.

REFERENCES

1. Hirayama K, Toyokura Y, Tsubaki T. Juvenile muscular atrophy of unilateral upper extremity: a new clinical entity. Psychiatr Neurol Jpn. 1959;61:2190–2197.
2. Hassan KM, Sahni H. Nosology of juvenile muscular atrophy of distal upper extremity: from monomelic amyotrophy to Hirayama disease—Indian perspective. Biomed Res Int. 2013;2013:478516.
3. Dejobert M, Geffray A, Delpierre C, Chassande B, Larrieu E, Magni C. Hirayama disease: three cases. Diagn Interv Imaging. 2013;94(3):319–323. doi:10.1016/j.diii.2012.10.008.
4. Boruah DK, Prakash A, Gogoi BB, Yadav RR, Dhingani DD, Sarma B. The importance of flexion MRI in Hirayama disease with special reference to laminodural space measurements. AJNR Am J Neuroradiol. 2018;39(5):974–980. doi:10.3174/ajnr.A5577.
5. Gomathy SB, Agarwal A, Garg A, Vishnu VY. Hirayama disease: review on pathophysiology, clinical features, diagnosis and treatment. touchREVIEWS Neurol. 2022;18(2):109–116. doi:10.17925/USN.2022.18.2.109.
6. Jin X, Jiang JY, Lu FZ, Xia XL, Wang LX, Zheng CJ. Electrophysiological differences between Hirayama disease, amyotrophic lateral sclerosis and cervical spondylotic amyotrophy. BMC Musculoskelet Disord. 2014;15:349.
7. Tokumaru Y, Hirayama K. Cervical collar therapy for juvenile muscular atrophy of distal upper extremity (Hirayama disease): results from 38 cases. Rinsho Shinkeigaku. 2001;41(4-5):173–178.
8. Gomathy SB, Priyanka Y, Garg A, Macken WL, Agarwal A, Ahmed T, et al. Phenotypic, electrophysiologic, and imaging spectrum of Hirayama disease from Northern India. Ann Indian Acad Neurol. 2024;27(5):524–529. doi:10.4103/aian.aian_348_24.